Radiation therapy for juvenile pilocytic astrocytoma of the pituitary stalk

Case report

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A case of juvenile pilocytic astrocytoma of the pituitary stalk is reported. The patient presented with diabetes insipidus and growth retardation. Magnetic resonance (MR) imaging was useful in diagnosing the lesion. Although the tumor could not be completely removed by surgery, radiotherapy was effective and the residual tumor disappeared 6 months after irradiation. The patient's condition has remained stable and no recurrence has been observed by MR imaging in the 4 years since radiation therapy.

Key Words • radiation therapy • juvenile pilocytic astrocytoma • pituitary tumor

The effectiveness of irradiation for treating juvenile pilocytic astrocytoma of the optic nerve or cerebellum is controversial. Some authors, mainly radiotherapists, have insisted on its efficacy, while others have disagreed. Even among the former group of authors, there is some controversy. Statistical analysis of a large number of such cases has shown that these tumors grow very slowly, characteristically decelerate gradually, then stop growing at some stage in their course, often irrespective of the use of irradiation.

Recently, we treated a small juvenile pilocytic astrocytoma of the pituitary stalk in a 6-year-old girl. The tumor completely disappeared 6 months after irradiation.

Case Report

This 6-year-old girl first presented with polyuria and polydipsia at the age of 3 years. Her urine output was 3500 to 4200 ml/day. A 4-hour water deprivation test produced mild urinary concentration (from 63 to 163 mOsm/liter), whereas intravenous Pitressin (vasopressin) administration caused remarkable concentration (from 58 to 487 mOsm/liter).

Examination. Computerized tomography (CT) scans and renal function were normal. Accordingly, the patient was diagnosed as having idiopathic diabetes insipidus and was treated with 1-desamino-8-D-arginine vasopressin (DDAVP). The diabetes insipidus was well controlled by DDAVP therapy, but the patient did not increase in height in a satisfactory manner.

Three years later, the patient was examined by magnetic resonance (MR) imaging, which clearly disclosed a tiny mass in the upper pituitary stalk and infundibulum. It appeared isointense relative to the optic nerve on T1-weighted MR images and hyperintense on proton density-weighted images (Fig. 1). Computerized tomography scans disclosed that the pituitary stalk was thicker than the basilar artery. The tentative diagnosis was either germinoma or astrocytoma.

Hormonal examination showed a growth hormone deficiency of hypothalamic origin. There was no response to insulin, levodopa, or arginine stimulation, but a response to growth hormone-releasing factor was noted. The base prolactin level was normal and a good response to thyroid-stimulating hormone was noted. Thyroid function was also normal. The cerebrospinal fluid (CSF) cell count was 1 to 2/cu mm and no abnormal cells were found. Beta-human chorionic gonadotropin was not detectable in CSF and blood samples.

Operation. The patient underwent tumor biopsy on June 24, 1987. The pituitary stalk showed a tumorous bulge from which a small biopsy was taken. Histological
findings were compatible with a juvenile pilocytic astrocytoma (Fig. 2).

Postoperative Course. The postoperative course was uneventful. The tumor site was irradiated using a rotational field with 10-MV x-rays from a linear accelerator. The total dose given was 45 Gy in 25 fractions. Follow-up MR imaging disclosed a gradual reduction in tumor size and finally complete disappearance of the tumor 6 months later (Fig. 3). The patient's condition has remained stable, but she is still receiving DDAVP. Furthermore, growth hormone replacement was started in August, 1988, to improve her growth retardation. She currently leads a normal life and no tumor recurrence has been revealed by MR imaging 4 years after irradiation.

Discussion

Infundibuloma

Glioma of the infundibulum or the pituitary stalk is a rare lesion, and reports of such tumors have been very few. Globus' called it "infundibuloma." He considered that the vascular structure of the infundibulum differed from that in other parts of the brain and that gliomas originating from the infundibulum also had this special vascular structure. However, infundibuloma is now no longer regarded as a special tumor but as a subgroup of astrocytoma. Actually, the histological findings of the two cases reported by Globus were compatible with those of pilocytic astrocytoma, which was the histological diagnosis in our case. Because of the age of the patient, the tumor was diagnosed as a juvenile pilocytic astrocytoma, a lesion that is commonly seen in the optic nerve or cerebellum.

Magnetic Resonance Imaging Characteristics of Pilocytic Astrocytoma

Reports of the MR imaging characteristics of juvenile pilocytic astrocytomas have been few. Lee, et al. reported that pilocytic astrocytomas appeared hypointense or isointense on T1-weighted images and hyperintense on T2-weighted images. The tumor in our patient was isointense on T1-weighted images and hyperintense on proton density-weighted images.

Optic Nerve Glioma

Zülch clearly stated that histologically optic nerve glioma is a pilocytic astrocytoma. Russell and Rubinstein expressed the same opinion, but they also admitted that some of these tumors include oligodendrocytic components. Zülch reported that pilocytic astrocytoma may occur at any site along the optic nerve and also in the hypothalamus.

In general, pilocytic astrocytomas are biologically the most benign type of glioma. Accordingly, when possible, surgical removal is the best choice of treatment, but these tumors cannot always be completely removed owing to their location, as in our case.

Optic nerve pilocytic astrocytomas are thought to have two biological forms, one being a very actively growing tumor and the other being a static lesion. To investigate this complicating phenomenon, Alvord and
Lofton collected and statistically analyzed 623 optic nerve gliomas from the literature and concluded that they grow rapidly at first but that eventually their growth decelerates in the natural course of the disease. Other authors have confirmed these results. Owing to this characteristic, the effect of irradiation on this tumor has been controversial.

Treatment Options for Optic Glioma

McFadzean, et al., reported on four cases of optic glioma treated with irradiation followed by CT, of which the lesion disappeared in three. Gunnesson-Nordin, et al., reported that five patients treated with an absorbed dose of 39 Gy or more were alive without signs of progression from 7 to 20 years after treatment. Danoff, et al., reported that the stabilization of visual impairment or an improvement in vision was noted in 78% of patients. Similar results were obtained by Dosoretz, et al., Harter, et al., Brand and Hoover, and Horwich and Bloom. Montgomery, et al., reported that radiotherapy was effective in preventing the progression of optic gliomas; they recommended treatment early in the course of the disease to minimize the associated visual deficit.

Only a few authors have used irradiation in selected cases. For example, Wallner, et al., recommended postoperative radiotherapy only for patients who had incomplete tumor resection, who were older than 3 years of age, and in whom tumor progression was documented by CT or MR imaging. Furthermore, Packer, et al., reported that the beneficial effects of radiotherapy were difficult to document.

Conclusions

In our patient, the tumor could not be completely removed and radiotherapy was given, achieving remarkable tumor regression. In this case at least, radiotherapy played an important role in the treatment of juvenile pilocytic astrocytoma since total resection was not possible.

References

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